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SHORT REPORT

Aortic Root Anomalies of the Neck Presenting in Adults. Review of the Literature with Three Case Reports

S.K. Das,* T.D. Brow and R. Byrom

Department of Vascular Surgery, The Hillingdon Hospital, Pield Heath Rd, Uxbridge, Middlesex UB8 3NN, UK

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Introduction

Anomalies of supra-aortic arterial anatomy sporadically challenge routine practice of vascular surgeons. The final anatomical configuration of the aortic root is closely linked to the embryological development of the aortic arch. The development of the aortic arch may produce numerous variations. Anomalous arch vessels can surround and compress the esophagus, and/or, trachea.[1] Gross was the first to divide a double aortic arch and termed them 'vascular rings'.[2]

Vascular surgeons may be required to manage anomalies of the aortic arch. Typically 'vascular rings' present to paediatricians in infancy, however, some may present in adult life. The majority of vascular rings that reach adulthood undetected remain asymptomatic. The commonest anomaly, sometimes referred to as a 'ring', is an aberrant right subclavian. It is present in .5% of individuals with more than 95% of aberrant right subclavian anomalies remaining asymptomatic.[3] Symptomatic cases are likely to require surgical intervention.

Anomalous vessels in the neck other than vascular rings may also have clinical relevance. Although the anatomy of the carotid tree remains remarkably constant, anomalous patterns can rarely arise. Right common carotid artery and subclavian artery may arise directly from the aortic arch with the absence of a brachiocephalic trunk. The common carotid artery may have a very low or high division. A thoracic carotid bifurcation was found in 2% of angiograms. These patients may present in different ways depending on the time of presentation and associated atherosclerotic disease. A lack of awareness and experience leading to an inability to deal with complex surgical anomalies may put the life of a patient at risk. It, therefore, remains vital that such surgical anomalies are discussed extensively and frequently in the literature. Discussion should focus on normal embryological development and their aberrations. With this objective we present three case reports and review the literature.

* Corresponding author. S.K. Das, MS, FRCS, MNAMS, M.Phil., Department of Surgery, Hillingdon Hospital NHS Trust, Pield Heath Road, Uxbridge, Middlesex UB8 3NN, UK.
E-mail address: saroj.das@imperial.ac.uk

Case 1

Symptomatic aberrant right subclavian artery (dysphagia lusoria)

A 64-year-old female presented to the gastroenterologists complaining of dysphagia, a choking sensation and weight loss. An endoscopy was normal. An ultrasound examination of the neck demonstrated a normal thyroid gland. A chest X-ray was normal with a left sided aortic arch. A CT scan was consistent with an aberrant right subclavian artery that arose as the most distal aortic branch and coursed behind the oesophagus and trachea in front of the vertebral bodies. A barium swallow, combined with an arch angiogram, revealed a posterior indentation of the oesophagus at the level of T4 by the abnormal right

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subclavian artery (Fig. 1). The right and left common carotid arose directly from the arch of the aorta.

Operation: transposition of the right subclavian artery to the right common carotid artery

Through a right supra-clavicular incision the prevertebral fascia on scalenus anterior was divided safeguarding the phrenic nerve. The carotid sheath was opened safeguarding the vagus. The internal and external jugular veins were ligated and divided. The right common carotid, subclavian, internal mammary, vertebral and costo-cervical trunk vessels were all identified. The aberrant subclavian artery was dissected behind the oesophagus to its origin from the aortic arch. The anomalous right subclavian was divided as far proximal to its origin and ligated. The distal end of the right subclavian artery was transposed to the right common carotid artery using a Javid shunt. Post operatively the patient made an uncomplicated recovery with relief of dysphagia at follow up.

Case 2

Right common carotid occlusion with an aberrant right subclavian artery

A 65-year-old left-handed male presented with a history of recurrent intermittent weakness in the left arm and left leg. A carotid duplex examination demonstrated a blocked right common and internal carotid artery with an 80% stenosis of the left internal carotid artery confirmed by arch aortography. The arch study further demonstrated an aberrant right subclavian artery arising distally with an occluded right common carotid artery arising directly from the aortic arch. A brain CT scan revealed minor infarcts in the region of the right parietal and right occipital lobes. Despite being on warfarin he had further transient ischaemic attacks thought attributable to both cerebral hemispheres. He underwent a left carotid endarterectomy. It is likely that he suffered from recurrent right carotid stump syndrome that was not controlled with anticoagulation.

Fig. 1. Barium swallow combined with aortography for case 1.

Case 3

Low carotid bifurcation

A 71-year-old male developed weakness in the right hand and a left sided visual field defect. A carotid duplex scan revealed 80% stenosis of the left internal carotid artery. No signal was demonstrated from the left external carotid artery. The right internal and external carotid arteries were normal on duplex. A left carotid endarterectomy was performed. At operation there was a normal internal and common carotid artery but no external carotid artery or external carotid artery branches identified. The ansa hypoglossi, vagus and hypoglossal nerves were normal. There was severe plaque at the usual site of origin of the internal carotid with subplaque haemorrhage. A routine carotid endarterectomy was performed. Post operatively he made an uneventful recovery. A follow up CT angiogram demonstrated that the bifurcation of the common carotid at 6 cm below the hyoid and the external carotid running normally in a plane parallel to the internal carotid.

Discussion

Arch embryology

Congdon showed that during the 4th and 5th week in the embryo the aortic arch system arises from the six primitive arch system connecting the aortic sac and the two dorsal aortas.[4] Though there are never more than four arches visible, a series of involutions and regressions affect the arch system. The first arch

becomes the mandibular artery, the second, the hyoid artery and the third, along with some cranial dorsal aorta, becomes the internal carotid. Meanwhile the heart and arches migrate caudally in relation to the neural tube, explaining the asymmetrical course of the recurrent laryngeal nerves.[5] At one point the fourth arch forms, in essence, a double aortic arch, this stage itself may be arrested, resulting in the first category of

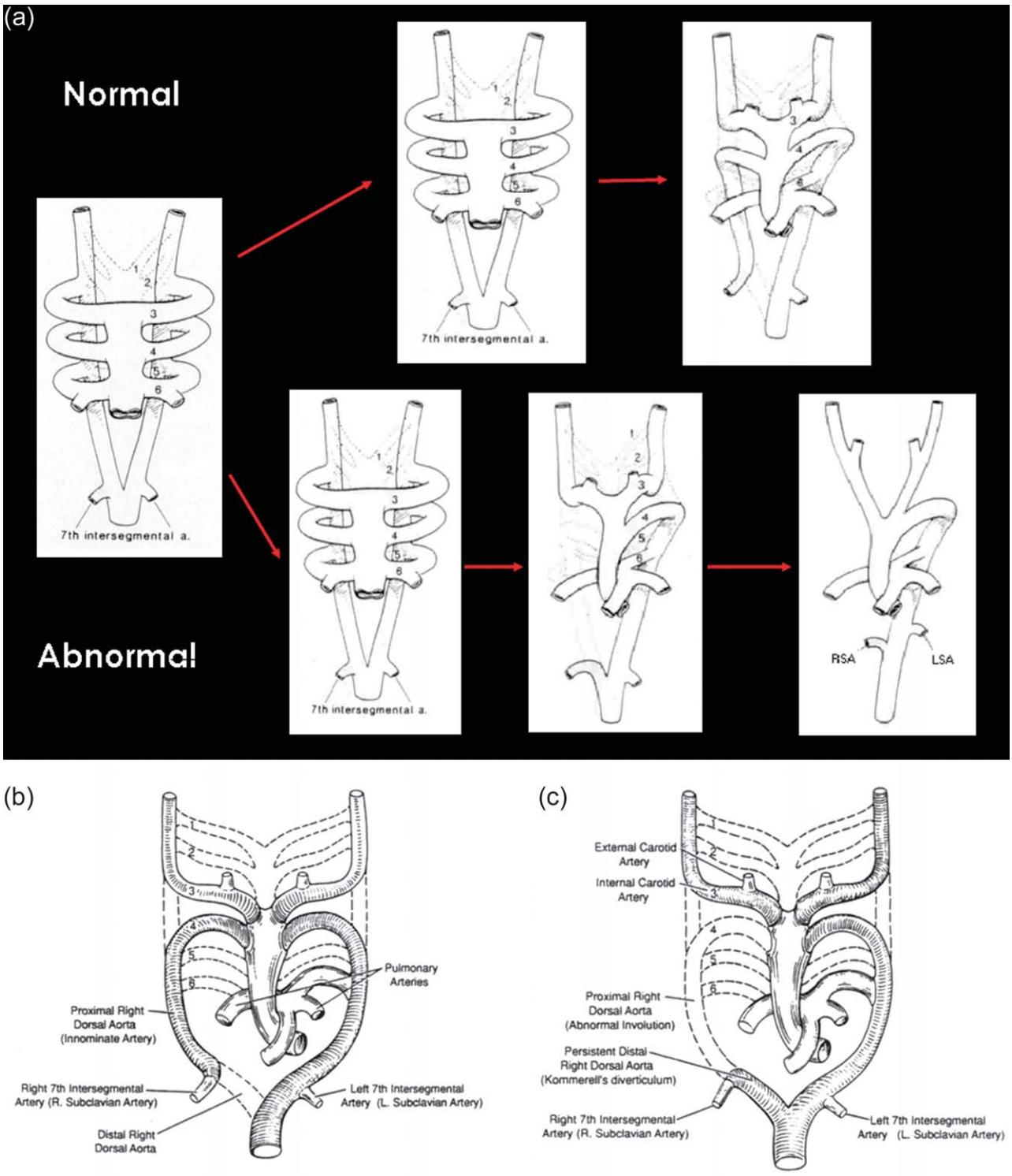

Fig. 2. (a) Normal and abnormal fourth arch development. (b) Normal embryological development with disintegration of the distal right dorsal aorta. (c) An aberrant right subclavian artery due to a persisting right dorsal aorta.

vascular ring. This stage formed the basis for the hypothetical double aortic arch plan of Edwards.[6] From this plan anomalies can be seen to develop according to the points of involution.[6–9]

Normally the right fourth arch joins the right seventh intersegmental artery and becomes the proximal segment of the right subclavian artery. In doing so the fourth right arch involutes leaving the left fourth arch to form the aortic segment between the left common carotid and the left subclavian artery[5] (Fig. 2(a–c)). The left seventh intersegmental artery forms the left subclavian artery. Aberrant right subclavian artery arises due to regression of the right proximal aortic segment and a persisting right distal eighth aortic segment, which normally disappears. (Fig. 2(b) and (c)) The right subclavian may arise from the eighth intersegmental artery, the seventh intersegmental artery joining this. When the aortic system is complete this becomes the last branch of the aorta and then extends to the arm.

Classification of aortic root anomalies

Aortic arch anomalies have been classified by various systems; some involving as many as 32 categories. There are five broad groups of aortic arch anomalies relevant to the vascular surgeons: (1) double aortic arch, (2) left aortic arch, (3) right aortic arch, (4) cervical aortic arch, (5) carotid anomalies.

Conclusion

The categorisation of four basic arch anomalies gives understanding to the term vascular rings and explains the types of aberrant right subclavian, i.e. occurring with a left arch or with a right arch. The carotid tree is considered separate from the arch anomalies. These anomalies clearly have important clinical implications not just in the approach to cannulation and surgery but also in recognition and imaging. Routine practice with minimal investigation may put patients at risk of missed anomalies. However, the cost-benefit and risks of intensive radiological screening have to be weighed against the unlikelihood of finding an anomaly. Aggressive imaging approach cannot be recommended routinely as the clinically significant anomalies are so rare. An awareness of these anomalies is important to the vascular surgeon not only in diagnosis and explaining the pathogenesis of symptoms but also while performing and planning procedures in order to avoid accidental injury.

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